

Coexistence of Extraskkeletal Mesenchymal Chondrosarcoma and Isolated Hemihyperplasia: A Case Report

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What to Learn from this Article?

The possibility of malignancy must be kept in mind in musculoskeletal masses of patients with hemihyperplasia.

Abstract

Introduction: Mesenchymal chondrosarcoma is a malignant tumor which originates from chondrogenic tissue. Aggressive surgery is considered the major treatment method for the primary disease but there may be unsatisfactory results of surgery depending on location or extension to unresectable sites of majority of mesenchymal chondrosarcomas. Hemihyperplasia, formerly called hemihypertrophy, is a rare congenital overgrowth disorder in which one or more body parts on one side of the body grows more than other, causing asymmetry. Isolated hemihyperplasia is caused by an underlying abnormality of cell proliferation in individuals with neither any other underlying diagnosis nor vascular or skin anomalies. Isolated hemihyperplasia reveals an increased risk of embryonal tumors such as Wilms tumor, adrenal carcinoma or hepatoblastoma. However no such concomitance has been reported with chondrosarcoma. We report the first case of mesenchymal chondrosarcoma presenting with isolated hemihypertrophy.

Case Report: A 64-year-old female presented with a palpable mass in posterolateral aspect of the left lateral malleolus which developed over 5 months. Hemihypertrophy involving the right side of her body was present and the patient had a history of foot shortening operation at adolescent age. Further examination of the patient revealed no vascular or skin anomalies, nor any findings suggesting a syndrome. Isolated hemihyperplasia diagnosis was achieved. After preoperative imaging studies, surgical excision was considered as best treatment option in local musculoskeletal tumor council and marginal resection was performed. The resected mass was sent for histopathological investigation. Immunohistochemical and morphological findings revealed malignant mesenchymal chondrosarcoma.

Conclusion: IH is well known to be associated with increased incidence of some embryonal tumors like Wilms' tumor, adrenal carcinoma and hepatoblastoma. But no such association has been reported with chondrosarcoma. In musculoskeletal masses of patients with hemihyperplasia, the possibility of malignancy must be kept in mind.

Keywords: chondrosarcoma, hemihyperplasia, mesenchymal tumor.

Introduction

Mesenchymal chondrosarcoma is a malignant tumor which originates from chondrogenic tissue. Aggressive surgery is considered the major treatment method for the primary disease but there may be unsatisfactory results of surgery depending on location or extension to unresectable sites of majority of mesenchymal chondrosarcomas. Hemihyperplasia, formerly called hemihypertrophy, is a rare congenital overgrowth

disorder in which one or more body parts on one side of the body grows more than other, causing asymmetry. Isolated hemihyperplasia is caused by an underlying abnormality of cell proliferation in individuals with neither any other underlying diagnosis nor vascular or skin anomalies. Isolated hemihyperplasia reveals an increased risk of embryonal tumors such as Wilms tumor, adrenal carcinoma or hepatoblastoma. However no such concomitance has been reported with chondrosarcoma. We report the first

Access this article online

Website:
www.jocr.co.in

DOI:
2250-0685.416

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Journal of Orthopaedic Case Reports | pISSN 2250-0685 | eISSN 2321-3817 | Available on www.jocr.co.in | doi:10.13107/jocr.2250-0685.416

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Figure 1:
Appearance of the left ankle. Note the heterogeneous palpable mass about 35 x 28 x 30 mm in posterolateral aspect of the left lateral malleolus.



Figure 2:
Hemihypertrophy involving the right side of body. Overgrowth of the right hand is obviously apparent.



Figure 3: Appearance of the lower extremities after left transtibial amputation. Notice the incision scar off foot shortening operation, the patient underwent at adolescent age.

case of mesenchymal chondrosarcoma presenting with isolated hemihypertrophy.

Case Report

We report a case of mesenchymal chondrosarcoma arising from the left ankle region of a patient with isolated hemihypertrophy of right side of her body.

A 64-year-old female presented with a palpable mass about 35 x 28 x 30 mm in posterolateral aspect of the left lateral malleolus which developed over 5 months (Fig. 1). Hemihypertrophy involving the right side of her body was present and the patient had a history of foot shortening operation at adolescent age. (Fig. 2, 3) Further examination of the patient revealed no vascular or skin anomalies, nor any findings suggesting a syndrome. Isolated hemihyperplasia diagnosis was achieved.

Imaging studies of the left ankle revealed; radiograph of the left ankle region was taken and it showed enlarged soft tissue shadow with large area of stipple calcification within it. From these findings diagnosis of the soft tissue sarcoma with calcification was kept. MRI revealed one heterogeneous hyperintense and hypointense large soft tissue mass lesion (Axial 36x31 mm, coronal 36x57 mm sized) in posterior left ankle region in T-2 and T-1 weighted images respectively. It was situated posterior to lateral malleolus adjacent to gastrocnemius muscle, between arteria tibialis posterior and achilles tendon. By contrast with accompanying soft tissue oedema, underlying bone was completely normal (Fig. 4).

Surgical excision was considered as best treatment option in local musculoskeletal tumor council and marginal resection was performed. Per-operatively mass and hematoma was observed (Fig. 5). Tumor had no osseous involvement with fibula. After dissection of tumor from adjacent tibialis posterior artery and achilles tendon, mass was excised. The resultant defect on achilles tendon was repaired with vicryl sutures. Following bleeding control, skin was closed with a flap from posterior calf. The resected mass was sent for histopathological investigation. Grossly, specimen was gray white colored and solid in consistency, containing local bleeding regions (Fig. 6). Microscopic examination showed immature cartilaginous tissue with osteoclast like giant cells and undifferentiated small round cells. The undifferentiated tumor cells were diffuse stained with Vimentin, CD99, BCL-2 and focally stained with actin. When immunohistochemical and morphological findings were considered together, diagnosis of malignant mesenchymal chondrosarcoma was achieved.

Postoperatively the patient was consulted with medical and radiation oncology and screening of the abdomen and chest were done which revealed evidence of pulmonary metastases. Chemotherapy was initiated. The patient presented surgical wound problems which turned out to be a local recurrence during chemotherapy (Fig. 7). Amputation of the left lower limb was

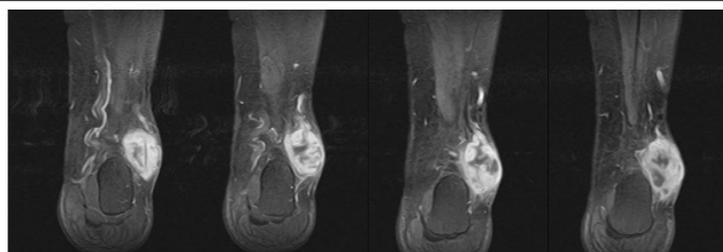


Figure 4: MRI clearly shows the heterogeneous large soft tissue mass lesion on the posterior aspect of left ankle.



Figure 5: Pre-operative view of the tumor and surrounding tissue. Infiltrated area of the skin was also excised with tumor.

performed. At last follow-up, the patient had multiple systemic metastases and severe respiratory distress.

Discussion

Chondrosarcoma is a malignant tumor of chondrogenic origin [1]. Four types of chondrosarcoma have been described: grade I, grade II, mesenchymal and myxoid [2]. The mesenchymal subtype is a rare and the most malignant variant of chondrosarcoma which is related with poor prognosis despite chemotherapy and radiotherapy [1, 3, 4, 5]. With a high incidence of local recurrence and distant metastasis, all cases presenting mesenchymal chondrosarcoma have been

reported fatal even after complete surgical excision [6]. The first description of mesenchymal chondrosarcoma (MC) in the literature was reported by Lichtenstein and Bernstein in 1954 [7, 8]. MC corresponds to less than 1% of all sarcomas and mostly affects children and young adults between the ages of 15 and 35 years [9]. Mesenchymal chondrosarcoma originates from chondrogenic tissue and is characterized by a bimorphic histological appearance, with both undifferentiated mesenchymal tissue and well-differentiated malignant hyaline cartilage [5, 8]. Most cases with MC are treated surgically although there have not been an agreement on a therapeutic standard, due to its very low incidence [9]. Aggressive surgery is considered the major treatment method for the primary disease but there may be unsatisfactory results of surgery depending on location or extension to unresectable sites of majority of mesenchymal chondrosarcomas [1].

Hemihyperplasia, formerly called hemihypertrophy, is a rare congenital overgrowth disorder in which one or more body parts on one side of the body grows more than other, causing asymmetry [10, 11, 12]. Hemihyperplasia may present in two forms: isolated or part of a syndrome. There have been several syndromes in which hemihyperplasia has been described, these include: Beckwith-Wiedemann syndrome, Proteus syndrome, Russell-Silver syndrome, Neurofibromatosis type I and Klippel-Trénaunay-Weber syndrome [10, 11]. Isolated hemihyperplasia is pragmatically described as asymmetric regional body overgrowth which is obviously apparent "from the end of the bed", and caused by an underlying abnormality of cell proliferation in individuals without any other underlying diagnosis [12]. Etiology of idiopathic nonsyndromic hemihypertrophy is unclear. There are no widely accepted diagnostic criteria to distinguish IH from normal growth variation. However absence of vascular or skin anomalies are interpreted in favor of IH as such anomalies are usually associated with the mentioned syndromes. Isolated hemihyperplasia reveals an increased risk of embryonal tumors such as Wilms tumor, adrenal carcinoma or hepatoblastoma [12]. Many prognostic implications and peculiar treatment options may be available for individuals with an underlying syndrome. Therefore before the diagnosis of IH is made, all the well known overgrowth syndromes with vascular or skin malformations should be ruled out preferably by a clinical geneticist experienced in the differentiation of IH from other causes of body asymmetry [10, 12, 13].



Figure 6: Macroscopic view of the tumor with local bleeding regions.



Figure 7: Appearance of the ankle after local recurrence, which lead to dehiscence of surgical area.

Sometimes the asymmetry or overgrowth of one side can be so slight that it isn't easily recognizable, and this leads to difficulties in determining the prevalence of isolated hemihyperplasia accurately. A study which included

[16].

In our case the patient was diagnosed as hemihyperplasia at adolescent age and had undergone foot shortening operation at that age. Since then the patient was neither called for orthopedics examination follow up nor screened for musculoskeletal tumors. At the age of 64 she presented with a palpable mass in posterolateral aspect of the left lateral malleolus which turned out to be a life threatening malignant tumor.

Conclusion

IH is well known to be associated with increased incidence of some embryonal tumors like Wilms' tumor, adrenal carcinoma and hepatoblastoma. But no such association has been reported with chondrosarcoma. In musculoskeletal masses of patients with hemihyperplasia, the possibility of malignancy must be kept in mind.

Clinical Message

It is reported that IH can coexist with embryonal tumors. However such a coexistence was not reported with chondrosarcoma until present case report. Chondrosarcoma is regarded to have different treatment and prognosis compared to embryonal tumors.

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Conflict of Interest: Nil
Source of Support: None

How to Cite this Article

Demirkiran ND, Akdeniz O, Hapa O, Havitcioglu H. Coexistence of Extraskeletal Mesenchymal Chondrosarcoma and Isolated Hemihyperplasia: A Case Report .*Journal of Orthopaedic Case Reports* 2016 April-June;6(2): 28-30